

Information for Secondary School Staff



Introduction

Prader-Willi syndrome (PWS) is a complex developmental disability that results from a defect on chromosome 15. It causes a malfunction in the area of the brain called the hypothalamus, which controls aspects such as appetite, temperature and emotion.

PWS can be characterized by:

- Hypotonia (low muscle tone)
- Hypogonadism (underdeveloped sex organs)
- Hyperphagia (uncontrollable hunger)
- Cognitive impairment
- Challenging behaviours

NB The combination of hypotonia and hyperphagia means that the child needs fewer calories than children of the same age to maintain a healthy weight.

Health professionals that may be involved with this child include:

- Dietician
- Endocrinologist
- Geneticist
- Occupational Therapist
- Ophthalmologist
- Orthopedic nurse or surgeon
- Orthoptist
- Orthotist
- Paediatrician
- Physiotherapist
- Speech therapist

(You may need to seek advice from the above in addition to reading this leaflet)



YouTube information video

You can view a 35 minute YouTube video which gives more information about children with PWS in a school setting at <https://www.youtube.com/watch?v=OxACKZRY6NQ>

A child with Prader-Willi syndrome is joining your school

This pack contains information which is appropriate to the age of this child and will help you to manage their needs, maintain a safe environment and facilitate learning.

But do remember - if you have any queries, we are here to help. We are only a phone call or email away.



What's special about this child?

Please remember that PWS is just a part of your pupil. They will have similarities and differences, as all other children do. Many people with PWS have specific and valuable attributes and skills. Many have excellent reading and writing skills as well as fine motor skills and are particularly clever with jigsaw puzzles and fine handiwork. They often have excellent long term memory and visual processing skills.

What to expect from your pupil and how you can best support them

Learning Disabilities and challenges

Many children with PWS experience some level of learning disability. The level of which varies from one child to the next. Please do be aware that even if the child presents as able, there may be underlying issues with regard to comprehension and cognitive ability. It is common for children with PWS to face challenges in expressive language use and short term memory.

What can you do to help?

Ensure instructions are repeated and ask the child to repeat instructions back to you. Check understanding and include 'refreshers' / revisit previous work regularly.

Dietary needs and food seeking behaviours

Food seeking behaviours are common in people with PWS and the pupil is likely to have lots of anxiety surrounding food. These types of behaviour are a result of the damage to the hypothalamus, meaning the child is unable to experience the sensation of fullness. It is very unlikely that a child with PWS would be able to make informed or safe decisions around food, particularly when left unattended.

What can you do to help?

It is important that all staff are aware and familiar with the child's dietary needs. It can sometimes be helpful to make other pupils aware of this too, providing you have parental permission. Anxiety can be reduced if food is kept out of sight and some form of food 'security' is in place. This might include keeping food storage areas locked, clearing empty plates away immediately after eating or ensuring lunch boxes are not left unattended. It is advised that you discuss any food within the classroom with the pupil's parents beforehand and that any food consumption for special occasions is arranged with them in advance. This can allow parents to 'borrow' calories from their evening meal. The pupil will need reassurance of his/her food routine which should include fixed times.



Emotional challenges

Some children with PWS have difficulty in controlling and understanding their emotions. You may find that emotional outbursts occur when the child feels anxious, frustrated, confused or that the situation is out of their control.

What can you do to help?

Ensure staff work together to reduce anxiety and to create positive and safe working environments. Reassure the child where possible and focus on achievements and goals.

Continued ...

What to expect from your pupil and how you can best support them (continued)

Anxiety

Most children with PWS suffer from some form of anxiety. Anxiety levels can increase when the child is faced with changes to their routine or when they are unable to predict the near future. At times, you may be able to see the child's anxiety growing with behaviours such as skin picking or fidgeting.

What can you do to help?

Ensure lines of communication between other staff and parents/carers are open in order to provide consistency and reassurance. Be clear about rules and routines and try not to deviate from these. If a change occurs that is unavoidable, inform the child as early as possible and be prepared to confirm these changes as often as needed.

Perseveration or repeated questioning

Repeated questioning is a common feature of PWS, which usually occurs when the child requires reassurance or if they have been unable to process the information given. On occasions, the child may use repeated questioning to gain 1:1 attention and verbal responses.

What can you do to help?

Agree to answer the question just 3 times. On the 3rd occasion, ask them to repeat your answer back and confirm they have understood your response.

If the child persists, try reducing verbal responses by shaking/nodding your head, or using visual reminders which they can refer to (eg. if the question is "what time is lunch?" try drawing a clock face on a paper plate to indicate your answer).

Time management

Some children with PWS struggle to move on from one activity to another, and you may find they are reluctant to move on until the task is completed. Most children with PWS are able to focus more effectively on several smaller tasks, as oppose to one large, on-going project.

What can you do to help?

Provide short activities and be clear on time frames or when you expect it to be completed. Ensure a gentle reminder is given as to how much time is remaining.

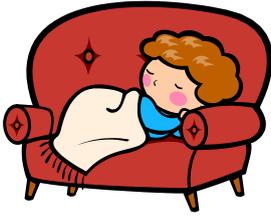
If the child insists they cannot move on until the task is complete, try issuing an "extension card", enabling them to borrow time which they can cash in later to complete the task.

Continued ...

Quick tips for behaviour management

- Use positive language
- Keep verbal responses to a minimum during behaviour incidents
- Use humour where possible
- Appear disinterested in negative behaviour
- Be consistent with your responses as a whole staff team
- Use a structured discipline procedure
- Work collaboratively with parents/carers

What to expect from your pupil and how you can best support them (continued)



Day time sleepiness

Note that children with PWS can often suffer from disturbed sleeping patterns. It is likely that this child will grow tired throughout the day and may require frequent rest breaks.

What can you do to help?

Ensure you have a quiet space available where the child can go to rest. It may be that the child requires a short nap (approx. 10 mins), and this may be required more than once a day.

Rewards

Positive behaviour should always be rewarded as children with PWS respond well to praise. They are also likely to enjoy responsibility and being told they've "done a good job". Doing this can boost the child's self-esteem and confidence.

What can you do to help?

Most importantly, food should **never** be used as a reward for a child with PWS. It can be useful to record achievements and praise on a graph or star chart. You can make this available for the child to refer to and the visual representation will make this accessible to them. When rewarding the whole class with food items, ensure you have consulted parents beforehand. They may provide a low calorie alternative or make arrangements for the calories to be borrowed from their evening meal.



We have a useful LEAPS Framework available on request , which includes further guidance for staff. Contact us for a copy

What can make behaviour worse?

A number of factors can make these behaviours worse, such as:

- Major changes in daily routine or environment
- Changes in diet or meal routine
- Conflicts of opinion amongst others in close relationship to the person with PWS
- Unexpected happenings or upsets
- Major life stresses such as bullying, teasing, bereavement, moving residence, changing schools etc
- Low mood

On a daily basis, very minor occurrences can affect behaviour. For example:

- Proximity to meal times
- Tiredness
- Minor change to routine
- Change in room or air temperature
- Mild teasing
- The teenager being unable to get their own way
- "Trigger" words or situations which are individual to the person



Medicines

It is likely that the child will take medication, usually in the form of a growth hormone (GH) injection. GH has positive effects in terms of muscle tone and height and is to be taken on a daily basis.

It is more than likely that this will be administered in the home, but if you are asked to keep medication on site please seek advice from the child's consultant or visit <https://www.gov.uk/government/publications/supporting-pupils-at-school-with-medical-conditions--3>

The right approach

The attitudes of other people are also very important.

Likely to have a **negative** effect on a teenager with PWS's behaviour are:

- Aggressive attitudes and tones of voice
- A confrontational approach
- Being overly sympathetic
- Displaying dislike of the person
- Not talking to the person
- Talking to the person too much
- Being in conflict with another person in authority - both giving different rules to the teenager with PWS, or arguing in front of him or her.
- Saying "Don't" as a means of trying to stop him doing something.

Likely to have a **positive** effect are:

- Firm, but loving and caring attitudes
- Ability to keep a sense of humour, and to see and exploit the funny side of a situation
- Ability to give clear guidelines and boundaries and stick to them
- Praise, and stressing the positive aspects of the person with PWS
- Saying "Do" as a way of showing him how to do something correctly, (ie instead of saying "Don't leave the door open", say "Please shut the door").

Your school environment

Many potential outbursts can be avoided by providing the right environment to suit your pupils needs. This might include:

- A structured day, with plenty of advance warning if changes are planned.
- Unpressurised tasks or expectations. For example, don't expect them to be ready to go out in five minutes if it normally takes them 20 minutes - give them plenty of warning, possibly using a "countdown" system, with a reminder at 15 minutes, then at 10 minutes, and so on. This is particularly important for those with higher intellectual levels who may be placed under greater pressure, because more is expected of them, but which they are often unable to deliver in the time space allowed. This includes educational tasks.

Continued ...

Your school environment (continued)



- Ensuring that all who come into contact with the pupil are giving the same message about what they expect in the way of behaviour.
 - Help them to learn techniques to keep calm such as deep breathing, listening to music, using worry beads or stress balls.
 - Put up notices and pictures to remind them to "keep calm" and/or of what happens when during the day.
- It may be useful to discuss in advance with him or her situations they find difficult and how to cope with the situation.
 - If the pupil finds it difficult to be around food (most, but not all, do), remove food from sight, do not allow others to eat in front of him or her and, if necessary, lock food away.
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Finding the right level of independence

Teenagers with PWS respond well to responsibility and the concept of Independence. It is likely that your pupil will be keen to learn independent living skills. However, it is worth bearing in mind that very few will have the capacity to live independently in adult life (most living at home or in residential or supported living arrangements).

Finding a level of independence whilst maintaining a safe environment can be a difficult balance, particularly around food. If the pupil takes part in a Life Skills course, try to focus the food preparation modules on Healthy Eating, or even Living with PWS. Speak to parents about their plans for the future, or what they hope for their son/daughter to achieve. This can avoid setting unrealistic expectations or setting the pupil up for disappointment later in life.

Supporting your pupil

Many children with PWS will have an Education, Health and Care plan. In addition to the support outlined in this, we would always recommend some form of support during social times and when around food to avoid high levels of anxiety or food seeking behaviours.

Children with PWS respond well to 1:1 support, and often this will be a requirement of their EHC plan. If you do already have support in place, do bear in mind that it can be very easy for people with PWS to become attached to this person, particularly if they are together for the entire school day. You may notice some obsessive behaviours surrounding this person. Whilst the consistency and reassurance of a familiar face can be beneficial, it is important to avoid the pupil taking 'ownership' of their support worker or TA. This can be easily overcome by the TA working with various pupils in the classroom, showing that he/she is not there solely for the pupil with PWS.

Continued ...

Supporting your pupil (continued)



It is common for children with PWS to be fond of adult company and the child may need some support in socialising with peers. Encouraging them to take part in lunchtime clubs or activities is beneficial in aiding their social development. You may find that the child will still require the support of a TA during these times, and it can often be useful to provide a quiet, 'safe' place if they become overwhelmed.

Sometimes a Positive Support programme can help develop social skills and relationship skills. You may notice the pupil struggles with conversational skills such as turn taking, waiting for others to finish speaking or listening to others. In cases such as these, it can be useful to play turn taking games such as board games or role playing games.

It is important to listen carefully to the parent(s) of the PWS child as they will understand their child's individual needs and idiosyncracies better than anyone else. It may appropriate to have more regular meetings with the parents than with other children in your class. A diary recording notable events in the day may also benefit both parent and teacher.

Special medical concerns

There can be health complications in PWS so it is important to be aware of warning signs that something is wrong.

If your pupil vomits it is imperative that this is taken seriously. People with PWS are unlikely to vomit at all and doing so **may** be a sign that something is wrong.

Due to their **high pain threshold**, any complaints of pain or discomfort should also be taken seriously. Most people with PWS will only complain of this if the pain is extreme, and even then the person may only describe it as an ache or discomfort.

It is important that all staff are aware that **bruising can occur easily**, even as a result of a slight knock or bump.

People with PWS also have **poor body thermostats** so the pupil may need extra support and guidance regarding appropriate dress.

Please do not assume that the child is able to attend to their own personal care or toileting. If you are unsure, his/her parents/carer will be able to direct you.

Information for Secondary School Staff

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